

# Ventilation and Duchenne



Frequently  
Asked  
Questions





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The information and advice in this booklet is to act as a general guide to ventilation, it should not be used as an endorsement of a particular treatment or to replace the services of your doctor or specialist. It should be used in conjunction with other information and medical advice from your doctor or specialist, who should be consulted in your health issues, particularly regarding any symptoms requiring diagnosis or treatment. Any action you take as a result of information given in this booklet is at your discretion. DMD Pathfinders cannot guarantee the accuracy of the information contained in this booklet, although we've made every effort to ensure it's correct. Any care or treatment being considered or received is particular to you as the individual.

Dr Jon Hastie and Mark Chapman both live with Duchenne Muscular Dystrophy (DMD). They are founder members of the charity, DMD Pathfinders, a user-led organization which promotes choice, control and quality of life for teenagers and adults with Duchenne, and are on the Steering Committee of Action Duchenne's Takin' Charge 14-19 Transition Programme for Young People with DMD.



In October 2013 Jon and Mark chaired an International Respiratory Meeting in London to discuss standards of respiratory care for adults living with DMD.

Speakers and delegates included Dr Jes Rahbek from Muskelvindfunden in Denmark, Professor Anita Simonds from the Royal Brompton Hospital, Dr Nicholas Hart and Angela Reddy from Lane Fox Respiratory Unit, Guys and St Thomas' Hospital, Dr Mark Elliott from Leeds, Dr Martin Allen from Stoke and Professor Francesco Muntoni from Great Ormond St Hospital.

The discussion focused on the need for high quality respiratory care services and ventilation to ensure a good quality of life for adults living with DMD.

*'I've used non-invasive ventilation since my early 20s, first at night, and now aged 33 I use it almost 24 hours a day. I chose not to have a tracheostomy as I'm managing well and non-invasive ventilation has fewer risks attached, although I would consider a tracheostomy if deemed necessary for my health. I find managing full-time NIV is easiest with a range of interfaces suited to each situation, allowing me to stay ventilated when eating, showering and dressing with minimal fuss. I use a combination of full-face mask, nasal pillows and mouthpiece ventilation. The mouthpiece is comfortable and non-intrusive and I like to use it when interacting with people as it seems less medical. Since using full-time NIV I feel healthier and more stable, with lots more energy - I feel great!' - Jon Hastie*



*'I began using non-invasive ventilation in my early 20s when it was being increasingly used on people affected by DMD who were experiencing respiratory issues. After a few years it was suggested I opt for 24-hour tracheostomy ventilation as my breathing issues were becoming more acute. Aged 26 I took the decision to go ahead, I'm now 44. Ventilation gave me a new lease of life, with increased energy, clearer speech, and much improved appetite. It also meant secretions could easily be cleared using suction through the tracheostomy, which significantly reduced chest infections. It has been surprisingly easy living with a tracheotomy. With the correct care and support I manage to live a fulfilled and active life. It was definitely the correct decision for me, giving a good quality of life for longer than I could have possibly imagined.'* – Mark Chapman



Jon and Mark presented evidence gathered from 300 members of DMD Pathfinders, this highlighted the differing practices and conflicting advice about best respiratory practice locally, nationally and internationally.

DMD Pathfinders has produced this Frequently Asked Questions (FAQ) booklet to address common questions and concerns on respiratory and ventilation issues.

Mark, aged 44, uses tracheostomy ventilation 24/7 and lives independently in Edinburgh with support from his team of Personal Assistants. Jon, aged 33, uses Non-invasive ventilation almost full-time and also lives independently in Shoreham-by-Sea supported by his team of Personal Assistants. Mark and Jon are experts by experience and contribute their knowledge and personal experiences of ventilation to this publication.

In order to express a preference for invasive or non-invasive ventilation, individuals living with DMD need access to current and relevant information.



## Starting Ventilation

### How do I know when I need ventilation?

If you suffer sleep disturbance, appetite-loss, have day-time fatigue, morning headaches, sleepiness during the day and concentration problems, or any combination of these, you are likely to be under-breathing at night and therefore in need of NIV.

NIV can also be required at a younger age to help resolve an acute chest infection or to support a person in the post-operative period, e.g. after scoliosis repair.



### How does ventilation support progress?

In most countries non-invasive ventilation is tried first when night-time use is required. In some countries there is a routine progression to tracheostomy ventilation when daytime ventilation is required. In other countries, including the UK, a combination of NIV, cough assistance and mouthpiece ventilation (sip) is used during the day.

### On average, when do people with DMD require ventilation support?

Between 18 to 20 years of age, but this can vary. Use of prednisolone (an oral steroid) in childhood may extend this by a few years. The introduction of ventilation support means that average survival now extends to the late 20s with many living into their 30s and 40s.

### What is a Respiratory (Ventilation) Clinic?



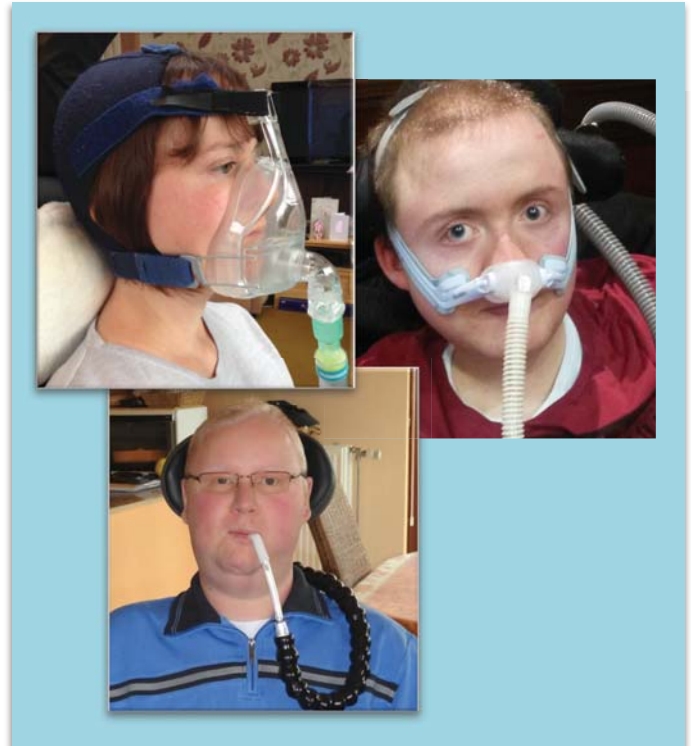
Once you start (don't wait!) to experience any of the symptoms of ventilation problems (see above, includes sleep disturbance, appetite loss), ask your GP/Consultant to refer you immediately to the nearest Respiratory Clinic. This should include a specialist doctor and therapist skilled in delivering NIV and other techniques for increasing the air entering the lungs, including manual and mechanically assisted coughing.

Where a person living with DMD needs ventilation, what are the different types of ventilation regimes?

There are two general types of ventilation used for people living with DMD:

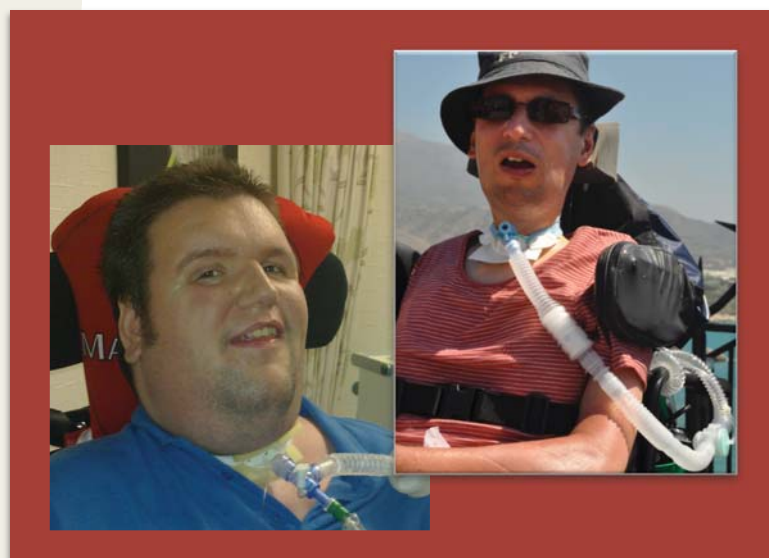
- Non-Invasive (NIV)

This is where the person with DMD will either wear a nasal interface, nasal mask or facemask or have a mouthpiece like a straw attached to their wheelchair (a SIP) through which ventilation is delivered, all of which are attached to a portable ventilator.



- Invasive

This is where a person with DMD has a surgical incision made in his windpipe (called a tracheostomy), through which a tube is inserted. Air goes in and out of the body through this tube instead of through the nose and mouth.



**Comparison table showing the main differences between  
Invasive and Non-Invasive Ventilation**

<b>Factor</b>	<b>NIV</b>	<b>Tracheostomy</b>
Risk when starting ventilation	No surgery is required to start non-invasive ventilation. There is a risk of pressure sores if the initial fit of the mask is not right.	All surgery carries a level of risk. An experienced surgical team can mitigate it but there will always be some risk of complications and (rarely) death.
Appearance	A SIP interface is less noticeable and sits in front of the mouth. A full facemask is highly visible and partially hides the face.	Some tracheostomy tubes can be hidden; others are highly visible
Coughing	DMD'er will be able to cough unaided or use manually or mechanically assisted coughing. Assisted coughing is more tiring than suctioning and may make colds and chest infections harder to deal with.	DMD'er will require coughing assistance and a suction machine to remove secretions from airways
Infection	No increased risk of infection if interface properly cleaned and maintained	There is an increased risk of infection
Speaking ability	Not much interference – depending on type of interface used. A full-face mask can muffle the voice and interfere with speech, while nasal pillows or SIP vents don't affect speech.	Ability to speak is altered but there are adaptations and adjustments that can help.
Swallowing	Doesn't interfere, aspiration may be more likely	May interfere, aspiration may be reduced slightly



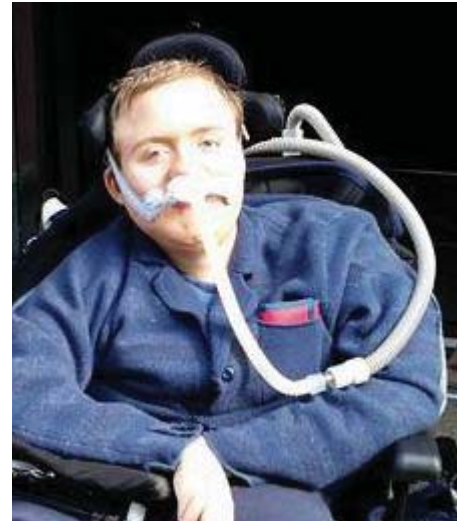
## What types of non-invasive ventilation are used?

There are two main types of non-invasive ventilation:

- Bi-PAP\* or bi-level ventilation

This type of ventilation uses air pressure to support your breathing. In pressure support mode, when you take a breath, this triggers the ventilator to use air pressure to ensure you inhale sufficient air to ensure your lungs inflate and then deflate properly. Ventilation can also be set so that the machine triggers at a certain number of times per minute and effectively takes over your breathing. Bi-level ventilation has both an inspiratory pressure, and a low expiratory pressure to enable breathing out.

*\*Bi-Pap refers to a particular make of ventilator*



- IPPV

IPPV stands for intermittent positive pressure ventilation. Like the BiPAP, it uses air pressure to support your breathing. However, unlike BiPAP, in IPPV there is only an inspiratory pressure, allowing your lungs to exhale on their own.

## What type of machine is used in non-invasive ventilation?

There are ranges of machines available that provide ventilation. Some simpler BiPAP machines may lack some of the more advanced features, while others (like the NIPPY 3) has a wide range of features and different ventilation modes. *When you need continuous breathing support, you will need a more sophisticated ventilator with some or all of these features:*



- A lightweight internal battery and the ability to auto transfer functions between power sources, an important safety feature
- Different modes or ways of triggering and delivering breaths
- Superior breath sensing and synchronizing capacity
- Multiple alarms to warn of a potential or impending problem

## Is CPAP used in ventilation (Continuous Positive Airway Pressure)?

No because it provides a constant pressure in inspiration and expiration (breathing in and out) it doesn't improve ventilation, therefore is not actually a form of ventilation. CPAP can be used in individuals with obstructive sleep apnoea but even here NIV is usually preferred.

## When should Invasive (tracheostomy) Ventilation be considered?

Invasive (Tracheostomy) Ventilation should be considered for some or all of these reasons:

- If respiratory failure is not controlled and NIV isn't working
- If aspiration occurs (when food is sucked into the airways), due to weak swallowing and ineffective coughing
- If there are problems clearing secretions which are not resolved with addition of physiotherapy and cough assist machine to NIV (see below)
- Personal preference

## How is a tracheostomy performed?

A tracheostomy is created by a surgical incision in the windpipe and without complications is relatively quick and straightforward to perform. It is usually performed under general anesthetic and like any surgical procedure with DMD this carries an element of risk that should be discussed beforehand with your doctor or consultant and the anaesthetist who'll be monitoring you during the operation. Recovering and dealing with the tracheostomy depends on a variety of factors such as: if it was an emergency or planned, your general health, the team's experience, how well it heals, if there's any infection, and the amount of aftercare required. The length of stay in hospital is determined by these factors as well as ensuring an adequate care package, that includes carer or PA training in tracheostomy management and suction, can meet your changed care needs at home.

## How do I maintain my tracheostomy?

The amount of care required to maintain a healthy tracheostomy does vary between individuals and can mainly be done at home. Some care is on a daily basis, like suctioning secretions (anything from 2 to 12 times per day, with or without a cough assist), cleaning leakages, and changing dressings, tracheostomy inner tubes and filters. Other care is less often, like a full tracheostomy tube change (every 1 to 4 weeks, but variable) and replacing the ventilator tubing circuit (at least every 3 months). Any changes in health like infections or having a cold will increase the amount of care and may occasionally result in short-term hospital admissions.



## Do I need specially trained nurses to deal with my tracheostomy?

Not necessarily, without any complications there is no reason carers or PAs cannot be trained to deal with tracheostomy care. It is extremely beneficial to receive back up support from a community nursing team, preferably specialising in ventilation, should any problems arise. They can also provide essential training, advice and guidance. Having a tracheostomy and being ventilator dependent it's important to have 24/7 care and support in place, with written plans covering common problems like tracheostomy tube blockage, tube displacement and chest infections.

## What help can I get with coughing?

Cough assistance techniques are used either alone or in combination with NIV or tracheostomy to move and clear secretions. Don't wait until your cough has become weak to get help because a good ability to cough is important in helping you clear secretions from your lungs and airways. A stepped approach is recommended – for some people physiotherapy/huffing (exhaling short, loud puffs of air) is sufficient, in others physiotherapy while using NIV is very helpful. It is also possible to provide big breaths to assist coughing via an ambu-bag or “self-inflating bag” (hand-held device used to manually provide positive pressure ventilation).



## What is a Cough Assist Machine?

If the above techniques are not sufficient to help with your coughing and you suffer from frequent chest infections, then a Cough assist machine is required (sometimes referred as a cough in-exsufflator). You should get one through your local respiratory clinic.



## What is a Suction Machine?

A Medical Suction Machine can be used to remove secretions from your lungs and airways, particularly with Invasive Ventilation. These machines use a vacuum pump and are often designed to be portable.



## Apart from equipment already mentioned, what else will I need?

You may need all or some of the following:

- Pulse Oximeter (1) – to monitor blood oxygen levels and pulse rate
- Humidifier (2) – delivers moisture through the ventilator tubing
- Nebuliser (3) – See information below
- Spare ventilator circuit (4) - ventilator tubing with exhalation valve
- Back-up ventilator – if you need the ventilator during the day as well as at night and can't breathe on your own for 4 hours or less.
- Spare battery/battery pack (6) - if you want to ventilate outside the home and for use during power cuts. Some ventilators have inbuilt batteries, but battery life may be relatively short. More information is available in DMD Pathfinder's separate factsheet: *'Life and Breath - Worry-free breathing during power failure'*: [dmdpathfinders.org.uk/advice-guides](http://dmdpathfinders.org.uk/advice-guides)



## What do I use a Nebuliser for?

Drugs for lung infections often have to be inhaled directly into the lungs. Not all DMD'ers can use an inhaler easily and a Nebuliser turns the medication into a fine mist that can be breathed into the lungs more easily than by inhaler, especially if your breathing-in muscles are weak.

Salbutamol (Ventolin) is most commonly nebulised. Salbutamol (Ventolin) opens the airways if the individual is wheezy and helps liquefy sticky secretions.

Steroids to reduce airway inflammation, and antibiotics can all be nebulised. Saline can also be nebulised to loosen thick phlegm.

## How do I get the equipment I need?

Get your neuromuscular specialist nurse, GP or consultant to refer you to the Respiratory/Ventilation Clinic where you will be assessed and receive help and training for the equipment that you require

## How will I feel when I start to need ventilation?

You might be nervous and self-conscious when meeting new people wearing whichever interface you use to deliver ventilation, e.g. a facemask. It's quite normal to feel like this! Why don't you contact someone with DMD who has been through this before you and who will listen to you and provide advice and support? – DMD Pathfinders provides a forum for adults living with Duchenne via its private Facebook group, to join please contact us: [www.dmdpathfinders.org.uk](http://www.dmdpathfinders.org.uk)

## Can I go out/travel with ventilation?



There is no reason for either invasive or non-invasive ventilation to interfere with this. Make sure you and your carers know what to do in an emergency. If out for longer periods you may need to take a portable suction unit or cough assist. Take a kit bag with you of supplies you might need, e.g. spare tubing, ventilator battery. Longer journeys and air travel are possible with careful prior planning involving you, your medical team, carers and airline companies.

Remember, life isn't over just because you need ventilation! DMD'ers do all kinds of things despite needing ventilation, e.g. study, travel, work, party.



## Will my sleep be affected by using ventilation?

It can be affected at first because it takes time:

- To get used to the breath rate
- To get comfortable with the mask positioning
- To find the ideal ventilation setting, headaches when waking up can be sign of inadequate ventilation, requiring an adjustment to the setting



## How do I eat when using ventilation?

It's important to make sure food and drink go down the right way! Some DMD'ers remove their NIV interface (e.g. mask) for eating. Others have to juggle eating and ventilator breathing. In this situation, timing is key and everyone has a different method that works for them, here's an example that works for a DMD Pathfinder with a tracheostomy:



1. Food in mouth during or just after first breath in
2. Wait until breath is out
3. Then swallow

## Will my wheelchair need to be adapted for ventilation?

Not usually. Ventilators have become small enough to attach easily to your wheelchair (much like you would attach a small rucksack).

If anything additional is required, your ventilation team or consultant should consult with wheelchair services or your wheelchair supplier.



## Are there other ventilator users I can contact?



### YES!!!

- DMD Pathfinders  
Join the private Facebook group for adults living with Duchenne or similar conditions. For more information please visit: [www.dmdpathfinders.org.uk](http://www.dmdpathfinders.org.uk) or email: [info@dmdpathfinders.org.uk](mailto:info@dmdpathfinders.org.uk)
- International Ventilator Users Network  
For more information visit: [www.ventusers.org](http://www.ventusers.org)

## What else can I do to take charge of my health?

Take sensible preventative measures, e.g. get the flu and pneumococcal vaccines, follow a good diet, have regular assessment of your nutrition needs, swallowing and heart functions, tell your friends and PAs/carers to stay away from you when they are ill!

## Is there a link between ventilation and cardiac function?



Yes, insufficient ventilation can cause increased stress on the heart. It is important to follow the advice of a neuromuscular respiratory specialist as to when to start ventilation. Delaying the use of ventilation against advice could cause cardiac problems.

## Is there a link between what I eat and ventilation?



Yes. Both malnutrition and obesity are harmful to respiratory function. Malnutrition badly affects respiratory muscles, reducing muscle strength. It additionally affects immune function, wound healing, invasive and non-invasive ventilator weaning (gradual removal of ventilation, e.g. after surgery), time off the ventilator and psychosocial function. Poor nutrition is associated with non-invasive ventilation failure.

Good nutrition is a critical part of long term DMD management, as is nutritional therapy, regular weight and nutritional intake monitoring and taking your time over meals.

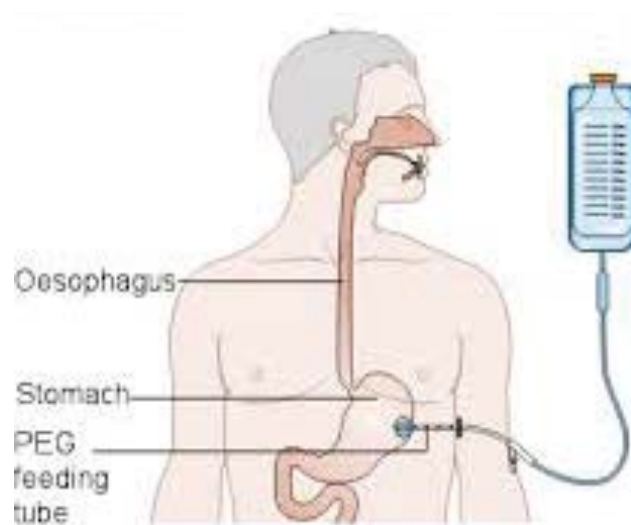
Common nutritional problems to watch out for:

- Weight loss or weight gain at different disease stages
- Chewing fatigue
- Dysphagia (difficulty in swallowing) and aspiration (food going down the wrong way)
- Inadequate nutritional intake
- Constipation
- Gastric dilatation (vomiting and pain caused by stomach and intestinal blockage)

## What is a PEG?

It's a feeding tube that is inserted through the stomach wall into the abdomen for the purpose of nutrition support. PEG is short for percutaneous endoscopic gastrostomy. PEG feeding is there to support normal eating and not necessarily to replace it, although it can, e.g. if you have severe problems with swallowing.

The PEG is also used to take medicines and additional water. Keeping hydrated is very important. PEG feeding can ensure adequate fibre (as well as hydration) and thus help relieve constipation. Your weight should increase providing more cover for your bones - thus making your wheelchair more comfortable.



## How can I keep myself healthy whilst living with DMD?

- Understand your condition!
- Talk to other DMD'ers about how to get the best physio/medical care - see [www.dmdpathfinders.org.uk](http://www.dmdpathfinders.org.uk)
- Talk to your Respiratory Clinic about breath stacking therapy. This therapy is performed using a lung volume recruitment device and consists of periodically filling your lungs to their maximum capacity, beyond what your weak muscles can achieve on their own
- Know the Standards of Care you are entitled to. Download a copy from [www.actionduchenne.org.uk](http://www.actionduchenne.org.uk)
- Monitor your condition so as to avoid crises
- Maintain good hygiene and make sure your carers do too, e.g. regular hand washing.





## What should I do in an emergency?

Remember, some health professionals and emergency staff may know nothing about DMD. YOU are the expert!

It's important to keep written information about how you should be treated by ambulance services and hospital staff. This should state the following:

- Your name.
- Your medical condition, e.g. DMD
- You are ventilator dependent.
- If you can breathe unaided and for how long (approx)
- How long your ventilator batteries will last (approx)
- You should not be given oxygen alone (unless there's a specific medical reason and it is used together with NIV ventilation).
- Your oxygen saturations should be monitored during transfer to hospital
- Equipment and supplies that must be taken with you, e.g. ventilator (including tubing circuit, mains power cable and batteries), cough assist, suction machine (including correct sized catheters), etc.
- If you require manual ventilation at any time with an ambu-bag they must ask you if they are bagging too hard/soft/fast/slow.
- Contact numbers of your respiratory consultant or specialist team (include their on call/emergency numbers).



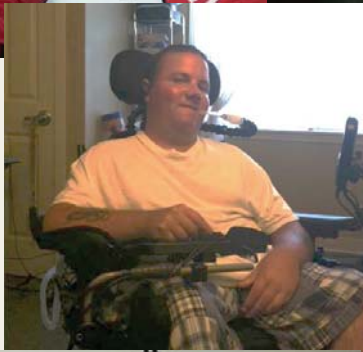
## What do I need to think about for the future?

- In order to receive a support package make sure you have a full assessment of your needs, this is your right and you should contact your local Social Services department, it could be that you will benefit from a personal health budget
- Know what you're entitled to, get good information, advice and guidance
- Talk to people in DMD Pathfinders who have been through this process
- Learn the skills to run your own life, e.g. through getting involved with Takin' Charge 14-19 Transition Programme for Young People living with DMD [www.actionduchenne.org.uk](http://www.actionduchenne.org.uk)





European statistics demonstrate that the introduction of ventilation in the management of Duchenne dramatically increases life expectancy. Members of DMD Pathfinders show us that there are many individuals using both invasive and non-invasive ventilation, who are relatively healthy and living productive, fulfilling lives. As an adult living with DMD, it is important that you have access to a specialist respiratory centre that can advise and support you in accessing ventilation at the appropriate time, using the method that best balances your clinical needs and personal preferences. It is extremely important that you do not delay introducing ventilation when it's needed, as this can have a knock-on effect on your heart, nutrition and general health. Hopefully this guide can help you to make the right choices and take control of your health, and will allow you to join an increasing number of vent users worldwide who are enjoying their lives



## *Thank you!*

DMD Pathfinders would like to thank all attendees of the International Respiratory Meeting, in particular Professor Anita Simonds (Royal Brompton Hospital, London), Dr Nicholas Hart and his team at the Lane Fox Respiratory Unit (Guys and St Thomas' Hospital, London), for their professional expert contributions in this booklet, Mark Chapman, Dr Jon Hastie and DMD Pathfinder members for their experiences and personal knowledge contributions. Thank you also to Celine Barry, Dr Janet Hoskins from Action Duchenne's Takin' Charge project and Action Duchenne themselves for making this publication happen.



DMD Pathfinders is run by and for adults living with Duchenne. It promotes choice and control and quality of life for teenagers and adults living with Duchenne Muscular Dystrophy in the UK. It campaigns for improved standards of health and social care and provides advice, guidance and support to teenagers and adults living with Duchenne.

DMD Pathfinders also provides a forum via our private Facebook group for adults with DMD or similar types of Muscular dystrophy.

Please consider donating to DMD Pathfinders as it's the only charity that can guarantee all funds go towards helping adults currently living with Duchenne. If you'd like to donate please visit the online fundraising page at [www.givey.com/dmdpathfinders](http://www.givey.com/dmdpathfinders)

If you would like to know more or join our private Facebook group for adults living with Duchenne, please visit our website [www.dmdpathfinders.org.uk](http://www.dmdpathfinders.org.uk) or email us at [info@dmdpathfinders.org.uk](mailto:info@dmdpathfinders.org.uk)